

Atrial Septal Defect With Right to Left Shunt Despite Normal Pulmonary Artery Pressure

JAMES D. THOMAS, MD, BURTON S. TABAKIN, MD, FACC, FRANK P. ITTLEMAN, MD

Burlington, Vermont

A 74 year old woman had right to left shunting through an atrial septal defect despite normal right heart pressures. Acute volume expansion temporarily reduced the shunt. Contrast echocardiography and angiography demonstrated that this shunting occurred almost exclusively from the inferior vena cava. At surgery a redun-

dant flap of septum secundum was found that was adjacent to the inferior vena cava orifice, intercepting its blood return like a spinnaker and shunting it into the left atrium.

(*J Am Coll Cardiol* 1987;9:221-4)

The occurrence of right to left shunting in patients with atrial septal defect is usually an ominous sign, heralding irreversible pulmonary hypertension. However, a small number of patients may have an anatomic anomaly that favors such shunting with normal right-sided pressures. We describe an elderly woman in whom a large eustachian valve and a redundant septum secundum caused preferential drainage of the inferior vena cava into the left atrium. We believe she is the oldest such patient to be described and displays an unusually variable degree of shunting.

Case Report

History. A 74 year old woman was admitted for evaluation of refractory hypoxemia. There was no history of congenital heart disease or heart murmur, and childhood growth and development were normal. She was quite active physically and had been an avid mountain climber. Approximately 4 years before admission, progressive exertional dyspnea developed. Evaluation at another hospital revealed an arterial partial pressure of oxygen (PO₂) of 41 mm Hg while the patient was breathing room air, rising to 50 mm Hg during breathing of 100% oxygen. A pulmonary artery catheter was placed; pulmonary capillary wedge pressure measured 2 mm Hg and pulmonary artery pressure 20/2 mm Hg. Volume infusion raised the pulmonary capillary

wedge pressure to 5 mm Hg and, coincident with this, raised arterial PO₂ to 120 mm Hg (while breathing 100% oxygen). She was transferred to this hospital for further evaluation. On questioning, she denied any knowledge of a heart murmur. She had stopped smoking cigarettes 10 years earlier after smoking 40 pack-years. She had had no occupational lung exposure and no significant pulmonary infection. There was a long-standing history of macrocytic anemia. Medication on transfer included isosorbide dinitrate and tocainide for unclear indications.

Physical examination. The patient was an elderly cachectic woman moderately dyspneic at rest and quite cyanotic. Blood pressure was 120/74 mm Hg with a heart rate of 80 beats/min and respirations 24/min. Positive physical findings were severe kyphosis with clear, though distant, breath sounds, very distant heart sounds without a murmur and a second heart sound that was virtually inaudible and without splitting.

Arterial blood gas determination while the patient was breathing 100% oxygen showed a pH of 7.47, a PO₂ of 59 mm Hg and a partial pressure of carbon dioxide (PCO₂) of 25 mm Hg. Electrolytes were normal and blood urea nitrogen was 19 mg/dl. Hematocrit was 36.0% (somewhat higher than usual), white blood cell count 11,900/mm³ and platelet count of 417,000/mm³. The electrocardiogram showed sinus rhythm at a heart rate of 80 beats/min with normal intervals and a QRS axis of 20°. Limb lead voltage was very low and there were only minor ST and T wave abnormalities and no atrial abnormality. Chest roentgenogram showed severe kyphosis with multiple vertebral compression fractures. Heart size was normal and lung vasculature and parenchyma were normal except for minor right lung atelectasis.

From the Departments of Cardiology and Cardiothoracic Surgery, Medical Center Hospital of Vermont, Burlington, Vermont.

Manuscript received March 10, 1986; revised manuscript received April 22, 1986, accepted May 14, 1986.

Address for reprints: James D. Thomas, MD, Massachusetts General Hospital, Non-Invasive Cardiac Laboratory, Boston, Massachusetts 02114.

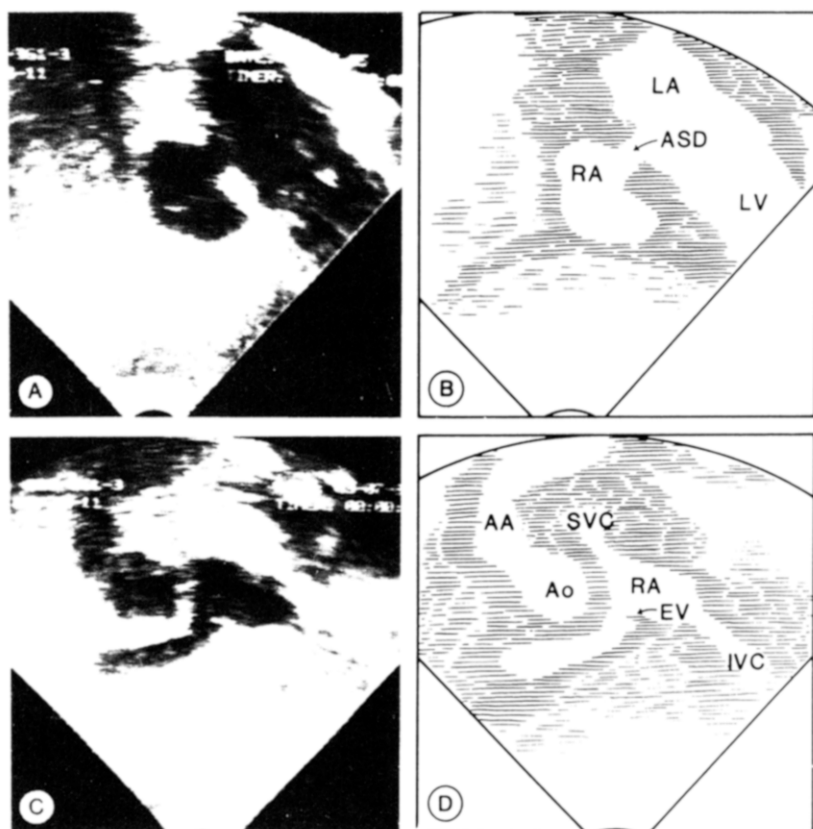


Figure 1. Echocardiograms demonstrating an atrial septal defect (ASD) (A and B) and large eustachian valve (EV) (C and D). Both frames are from the subcostal long-axis window. C and D also demonstrate striking distortion of the right atrium (RA) by the enlarged and ectatic aorta (Ao). AA = ascending aorta; IVC = inferior vena cava; LA = left atrium; SVC = superior vena cava.

Diagnostic studies. Evaluation for hypoxemia revealed the following test results. Vital capacity was 1.65 liters (62% of predicted) and forced expiratory volume at 1 second was 1.18 liters (62% of predicted). Radionuclide ventriculography (performed from the right arm) displayed normal ventricular function, a left ventricular ejection fraction of 60% and no evidence for an intracardiac shunt. Pulmonary perfusion scan (again from the right arm) showed minor lung irregularities and no systemic shunting. However, injection of technetium-tagged microspheres into the left foot demonstrated a significant right to left shunt. Echocardiography revealed a large atrial septal defect (Fig. 1A and B). The aortic root was enlarged, compressing and distorting the right atrium. A large eustachian valve was demonstrated in the inferior vena cava (Fig. 1C and D). Saline microbubble infusion showed normal drainage into the right atrium from the left and right arms and the left leg. However, significant right to left shunting was demonstrated at the atrial level, much more prominently from the inferior than from the superior vena cava. Left and right ventricular function appeared normal and the interventricular septum conformed to the left ventricle.

Cardiac catheterization was performed through the right femoral artery and vein. A flow-directed catheter was easily passed through the atrial septal defect. Oxygen saturation, measured while the patient was breathing 100% oxygen, and hemodynamic data are displayed in Table 1. It was

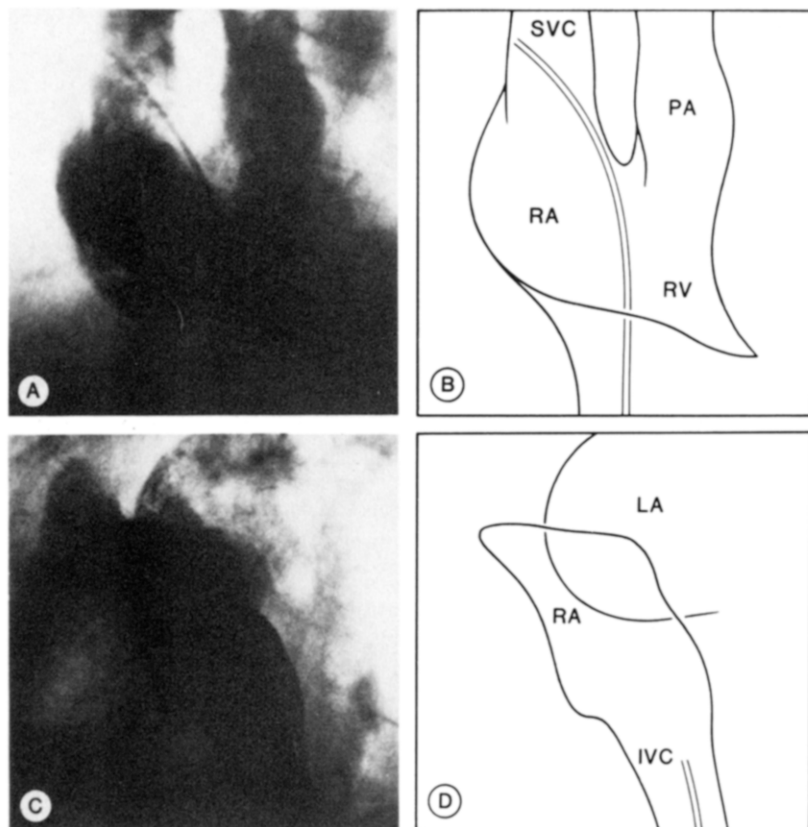
calculated that 33% of systemic venous return was shunting through the atrial septal defect with no detectable left to right shunt. The right-sided pressures were entirely normal and mean right atrial pressure was about 1 mm Hg less than left atrial pressure. The inferior vena cavogram (Fig. 2C and D) showed significant right to left interatrial flow, whereas the superior vena cavogram (Fig. 2A and B) showed almost no shunting. A left ventriculogram and coronary arteriograms were normal.

Surgery. It was elected to proceed with operative closure of the atrial septal defect. In the operating room, before the induction of anesthesia and fluid infusion, the patient's arterial PO_2 while breathing 100% oxygen was 50 mm Hg.

Table 1. Catheterization Data

Location	Pressure (mm Hg) (mean)	O ₂ Saturation (%)
Superior vena cava		70
Inferior vena cava		72
Right atrium	6/4 (2)	69
Right ventricle	15/2	70.5
Main pulmonary artery	15/4 (6)	70
Right pulmonary artery		70
Left lower pulmonary vein	(3)	100
Left atrium	7/4 (3)	90
Left ventricle	95/4	90
Aorta	95/55	90

Figure 2. Right atriograms with contrast injection into the superior vena cava (SVC) (A and B) and inferior vena cava (IVC) (C and D). Superior vena cava injection opacifies the right atrium (RA) and ventricle (RV) exclusively. With inferior vena cava injection there is opacification of the left atrium (LA) due to preferential shunting of blood across the atrial septal defect. PA = pulmonary artery.



After intubation and the infusion of 3 liters of Ringer's lactate solution, arterial PO_2 increased to 303 mm Hg.

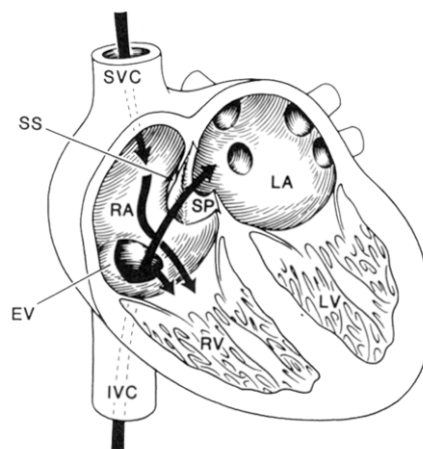
Before cannulation a purse-string suture was placed in the right atrial appendage and digital examination of the right atrium was undertaken. The septum appeared to be intact, except for a 1 to 1.5 cm defect located in the inferior aspect of the atrial septum adjacent to the inferior vena cava. The inferior vena cava was also explored and found to be essentially normal. Standard cardiopulmonary bypass was then established and myocardial preservation was achieved with the infusion of cold cardioplegic solution through the aortic root. The right atrium was opened and again the 1 to 1.5 cm defect was noted in the inferior aspect of the atrial septum adjacent to the opening of the inferior vena cava. There was a redundant septum secundum partially overlying this defect which could easily have a spinnaker effect depending on atrial pressures. The foramen ovale was closed, primarily incorporating the septum secundum in the closure. The patient was separated from bypass without difficulty and her postoperative convalescence was complicated only by a transient episode of congestive failure which resolved with diuretic drug therapy.

After extubation and adequate diuresis, the patient's rest PO_2 without supplemental oxygen therapy was 80 mm Hg. Clinically the cyanosis cleared, and subjectively she was much less dyspneic than she had been before surgery. She was discharged in good condition.

Discussion

Hemodynamics of atrial septal defect. In the absence of pulmonary hypertension, the typical shunting in atrial septal defect is overwhelmingly left to right (1-4). Careful

Figure 3. Schematic diagram of spinnaker formation from the redundant septum secundum (SS) allowing preferential shunting of inferior vena cava (IVC) blood into the left atrium (LA). With volume expansion, the interatrial septum was stretched, flattening out the "spinnaker" and reducing the right to left shunt. Note the prominent eustachian valve (EV) that helped to direct inferior vena cava blood onto the interatrial septum. LV = left ventricle; SP = septum primum; other abbreviations as in Figure 1.



pressure-flow studies have shown a small ($< 10\%$) right to left shunt in simple atrial septal defects occurring during brief periods of a right to left pressure gradient during the cardiac cycles. This is said to occur either early in atrial systole (the right atrium being activated before the left) (5) or during early ventricular systole (2).

Previous cases of right to left shunting without pulmonary hypertension. Only rarely has predominant right to left shunting at the atrial level been reported with normal right-sided pressures. Gallaher et al. (6) reported three patients, aged 26, 14 and 8 years, in whom a large eustachian valve directed inferior vena cava inflow through a secundum atrial septal defect (two patients) or patent foramen ovale (one patient) into the left atrium. Winters et al. (7) described a 52 year old man in whom right to left shunting through a previously undiagnosed atrial septal defect developed due to rotation of the heart after pneumonectomy. The anatomic shift caused the inferior vena cava to empty onto the atrial septum. The magnitude of the shunt in that patient had a marked positional variation; his arterial oxygen saturation fell from 91.6 to 75% when he changed from a lying to a sitting position.

Hausdorf et al. (8) recently described a case of cor triatriatum dexter in which a large eustachian valve covered the tricuspid valve orifice and shunted all systemic venous return into the left atrium. They also reviewed several examples of cor triatriatum dexter in which only the inferior vena cava drainage was shunted to the left with superior vena cava and coronary sinus return emptying normally into the right ventricle. This has been designated type III cor triatriatum dexter by Doucette and Knoblich (9). An interesting example of spinnaker formation of the sinus venosus valve was described by Jones and Niles (10). Depending on the direction of billowing, this valve caused obstruction of either systemic venous return or coronary sinus return, ultimately causing death in a 10 year old boy.

Etiology. The present case was marked by the late development of crippling hypoxemia due to preferential shunting of inferior vena cava blood into the left atrium. Two anatomic anomalies contributed to this shunting (Fig. 3). The first was a large eustachian valve that directed inferior vena cava flow to the interatrial septum; the second was a redundant septum secundum that billowed outward into the right atrium, intercepting inferior vena cava blood, and shunting it through the widely patent foramen ovale into the left atrium. This was not associated with high right-sided pressures; indeed, the mean right atrial pressure was about 1 mm Hg less than mean left atrial pressure. Of special interest in this case was the variable degree of shunting observed. On two separate occasions, it was documented that volume loading greatly alleviated the patient's cyanosis. A partial explanation may be that this increased left atrial pressure more than right. More importantly, it is likely that atrial dilation caused by volume infusion changed the anatomy of the atrial septum, decreasing the redundancy of the

septum secundum and allowing it to cover the margin of the septum primum, thus obliterating the foramen ovale. This observation may even explain the development of this lesion so late in life. Perhaps atrial distortion and reduction, due to the severe kyphosis, aortic root enlargement or even nitrate therapy, caused the septum secundum to fall away from the septum primum, billowing out into a spinnaker to intercept inferior vena cava drainage and direct it toward the opened foramen ovale. Such a situation would behave functionally like cor triatriatum dexter, type III. Simple closure of the foramen ovale with the septum secundum has relieved her hypoxemia.

Conclusions. Several points from this case deserve special emphasis. First, in assessing the site of right to left shunting, contrast echocardiography or radionuclide studies should be performed from the right arm, the left arm (looking for persistent left superior vena cava emptying into the left atrium) and the legs. Second, the occurrence of right to left shunting at the atrial level does not necessarily indicate severe pulmonary hypertension. The physical examination, electrocardiogram, chest roentgenogram and echocardiogram all suggested normal right heart pressures, confirmed by catheterization. Third, although surgery is the definitive solution in a case like this, some amelioration of the cyanosis may be obtained by changing the volume status or position of the patient.

We gratefully acknowledge the technical assistance of Joelyn Niggel in performing the echocardiographic examination on this patient.

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